PROC (BAYL UNIV MED CENT)
2022;35(1):58–59

https://doi.org/10.1080/08998280.2021.1960123

Taylor & Francis
Taylor & Francis Group

Management of asymptomatic severe hypertriglyceridemia

Nathalie V. Scherer, BS[a], and Dipesh Bista, MD[b]

[a]School of Medicine, Texas Christian University and University of North Texas Health Science Center, Fort Worth, Texas; [b]Department of Internal Medicine, Baylor Scott & White All Saints Medical Center, Fort Worth, Texas

ABSTRACT

Severe hypertriglyceridemia is an urgent presentation that requires acute treatment. We present a rare case that could not be controlled by medical management and required plasmapheresis.

KEYWORDS Plasmapheresis; severe hypertriglyceridemia; therapeutic plasma exchange; triglycerides

Triglycerides (TG) are lipids taken up from the digestive tract that are transported in plasma by lipoproteins and mobilized for energy through lipolysis.[1] Lipoprotein lipase is the primary enzyme of TG lipolysis and hydrolyzes TG to form fatty acids. Most cases of hypertriglyceridemia have a polygenic basis. There are also nongenetic determinants that can exacerbate hypertriglyceridemia, including obesity, metabolic syndrome, poorly controlled diabetes mellitus, pregnancy, and certain medications.[2] Severe hypertriglyceridemia is commonly associated with acute pancreatitis but can rarely cause multiorgan system dysfunction related to hyperviscosity, such as acute kidney injury and acute respiratory failure.[3]

CASE DESCRIPTION

A recently pregnant 33-year-old woman with known mild hyperlipidemia presented to an emergency department with fatigue, polyuria, polydipsia, and new onset yellow-pigmented papules on her chest and back. Initial workup showed blood glucose >1000 mg/dL, total cholesterol >900 mg/dL, TG 2788 mg/dL, high-density lipoprotein cholesterol 78 mg/dL, and low-density lipoprotein cholesterol 483 mg/dL. The patient was admitted for a hyperglycemic hyperosmolar state and started on an intravenous insulin drip along with intravenous hydration. Overnight, as her blood glucose was controlled, the intravenous insulin drip was stopped and transitioned to subcutaneous insulin. The next morning, however, her blood TG level was significantly elevated at 13,800 mg/dL. She was restarted on an intravenous insulin drip for acute management of severe hypertriglyceridemia. Repeat laboratory tests showed that blood TG had further increased to 20,100 mg/dL. A venous blood sample was notably milky and light pink. The patient did not show any evidence of acute pancreatitis but did have an elevated lactic acid level, which was concerning for impending end-organ dysfunction due to blood hyperviscosity.[3] Despite being on an insulin drip, fenofibrate, and omega-3 fatty acid, she had persistent, severe hypertriglyceridemia.

The patient was then sent for an urgent therapeutic plasma exchange (TPE) while continuing with the medication regimen. The plasma extracted was a thick, milky white color *(Figure 1)*, and the patient's TG dropped to 8406 mg/dL the following morning. The next 2 days, the patient continued to have daily TPE while being continued on all medications. The TPE was stopped after three rounds, when the patient's plasma TG dropped to 480 mg/dL on hospital day 5. During these 5 days, the patient had no abdominal pain or other clinical symptoms, although the yellow-pigmented papules persisted, presumed to be eruptive xanthelasmas associated with high TG.[4]

DISCUSSION

Patients presenting with hypertriglyceridemia of this severity are rare. When these patients do present, the highest priority is to lower TG as fast as possible to prevent morbidity and mortality that results from end-organ damage, which is associated with the increased viscosity of TG-rich plasma. This end-organ damage includes acute pancreatitis, acute kidney injury, acute respiratory failure, and

Corresponding author: Nathalie V. Scherer, BS, TCU and UNTHSC School of Medicine, TCU Box 297085, Fort Worth, TX 76129 (e-mail: n.v.scherer@tcu.edu)

The authors report no conflicts of interest. All involved persons gave their informed consent for this case report publication.
Received April 23, 2021; Revised July 18, 2021; Accepted July 22, 2021.

Figure 1. The first plasmapheresis with plasma that was thick and milky white in color.

myocardial infarction.[3] Intravenous insulin, fibrates, and omega-3 fatty acids decrease serum TG breakdown by upregulating lipoprotein lipase activity and reducing the production of new TG.[5] In patients with severe hypertriglyceridemia, information on the success of treatments has come primarily from case reports and retrospective chart reviews that have shown efficacy with the use of plasmapheresis.[6] TPE was indicated in our patient and was successful in reducing her severe hypertriglyceridemia.[7]

Chylomicronemia syndrome is defined as TG >1000 mg/dL with additional manifestations, such as eruptive xanthelasmas or lipemia retinalis.[8] Our patient met these criteria. A variety of underlying pathophysiological changes can give rise to chylomicronemia syndrome, but the precise etiology in our patient is unclear. The most common cause of chylomicronemia syndrome is familial hypertriglyceridemia, an autosomal dominant trait, but the patient had no relevant family history, and genetic testing was not indicated.[8] A variety of other factors can aid in developing this syndrome, such as uncontrolled diabetes mellitus, a diet high in fats, and pregnancy.[3] Given that this patient had given birth several months prior, presented with hyperosmolar hyperglycemia syndrome, and had a body mass index $\geq$25 kg/m^2, it is likely that her chylomicronemia syndrome developed from a combination of genetic predisposition and some modifiable risk factors.

ACKNOWLEDGMENTS

The authors would like to thank Dr. Mike Bernas, associate professor at TCU and UNTHSC School of Medicine, for his help with this manuscript.

1. Sandhofer F. Physiology and pathophysiology of the metabolism of lipoproteins. *Wien Med Wochenschr*. 1994;144(12–13):286–290.
2. Xiao C, Dash S, Morgantini C, Hegele RA, Lewis GF. Pharmacological targeting of the atherogenic dyslipidemia complex: the next frontier in CVD prevention beyond lowering LDL cholesterol. *Diabetes*. 2016;65(7):1767–1778. doi:10.2337/db16-0046.
3. Argula RG, Strange C, Budisavljevic MN. Multiorgan system dysfunction in the chylomicronemia syndrome. *J Intensive Care Med*. 2014; 29(3):175–178. doi:10.1177/0885066613475425.
4. Lee SY, Sheth CA. Eruptive xanthoma associated with severe hypertriglyceridemia and poorly controlled type 1 diabetes mellitus. *J Community Hosp Intern Med Perspect*. 2019;9(4):344–346. doi:10.1080/20009666.2019.1650591.
5. Lloret Linares C, Pelletier AL, Czernichow S, et al. Acute pancreatitis in a cohort of 129 patients referred for severe hypertriglyceridemia. *Pancreas*. 2008;37(1):13-2. doi:10.1097/MPA.0b013e31816074a1.
6. Stefanutti C, Di Giacomo S, Vivenzio A, et al. Therapeutic plasma exchange in patients with severe hypertriglyceridemia: a multicenter study: plasma exchange for hypertriglyceridemia. *Artif Organs*. 2009; 33(12):1096–1102. doi:10.1111/j.1525-1594.2009.00810.x.
7. Gupta N, Ahmed S, Shaffer L, Cavens P, Blankstein J. Severe hypertriglyceridemia induced pancreatitis in pregnancy. *Case Rep Obstet Gynecol*. 2014;2014:1–5. doi:10.1155/2014/485493.
8. Leaf DA. Chylomicronemia and the chylomicronemia syndrome: a practical approach to management. *Am J Med*. 2008;121(1):10–12. doi:10.1016/j.amjmed.2007.10.004.